

353

Is BNP level still correlated to echocardiographic indices in End-stage renal disease children on maintenance hemodialysis?

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Purpose: This study was designed to investigate the validity of brain natriuretic peptide (BNP) levels for the estimation of the systolo-diastolic ventricular function, and to determine the relationship between BNP levels and echocardiographic parameters of right heart chambers

Methods: We measured BNP levels and performed Doppler echocardiography in 35 end-stage renal disease (ESRD) children before (pre HD) and after hemodialysis (post HD). The relationships between BNP levels and 'conventional' and 'tissue Doppler' echocardiography indices were evaluated.

Results: Plasma BNP concentration increased significantly ($r=-0.41$, $p=0.018$) with decreasing left ventricular ejection fraction. Pre-HD BNP level was positively correlated to E deceleration time ($r=0.374$; $p=0.042$) and to peak pulmonary vein systolic velocity (S) ($r=0.405$, $p=0.019$). Before hemodialysis, the BNP level was significantly higher in children with early transmural LV filling velocity E/ LV lateral wall Ea >5 than those with E/Ea <5 ($p=0.04$). When E/Ea >5 and Ea/Aa <1 at the free wall tricuspid annulus, were combined in the same patient, the BNP concentration was significantly higher than those without these parameters ($p=0.006$). Post BNP concentration was positively correlated to LV end-systolic volume (LVESV) ($r=0.401$, $p=0.023$), to peak pulmonary vein (PV) systolic velocity (S) ($r=0.49$, $p=0.004$), to S/D ratio ($r=0.605$, $p<0.0001$), to Ea/Aa ratio measured at lateral mitral annulus ($r=0.453$, $p=0.008$), and RV Tei index ($r=0.389$, $p=0.037$).

Conclusion: This study shows that there is a significant correlation between echocardiographic parameters and concentrations of BNP in children. These data demonstrate how BNP may play an adjunctive role to echocardiography in assessing ESRD children.

354

Evolution of right ventricle volumes and function from childhood to adulthood

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Background: Progressive pulmonary regurgitation (PR) is a common complication after total surgical correction of tetralogy of Fallot (TOF). Long-standing PR leads to right ventricular (RV) dilation, which in turn causes RV dysfunction. Kinetics of RV dilatation during follow-up of these patients is unknown. We aimed to determine the rate of RV dilatation in repaired TOF, and the risk factors of RV dilatation

Method: Successive Cardiac Magnetic Resonances (CMR) was performed during 4 years in 64 patients with repaired TOF, aged 4-17 years (mean age $=17.3 \pm 8.2$ years).

Results: 56% ($n=36$) of patients were children (still growing). The surgical correction was performed at the median age of 5 months (1 month to 16 years). In the global population, the rate of RV end-diastolic (ED) volume dilatation was 0.7 ± 7.9 ml/m²/year, the rate of RV end-systolic (ES) volume dilatation was 0.3 ± 8.5 ml/m²/years. There was no difference between the children and the adults group. Dilatation of RV ED volume was significantly accelerated by the use of an infundibular patch for the surgical correction ($p=0.04$). Residual pulmonary stenosis, associated with PR, helps to maintain RV ejection fraction during follow-up ($p=0.019$). Age at surgery, history of palliated shunt, RV fibrosis had not influence on RV volumes kinetics.

Conclusion: RV dilatation may occurs very soon after surgical correction of TOF, then RV volumes were relatively stable during follow-up. However,

use of infundibular patch promotes RV dilatation and residual pulmonary stenosis could preserve RV systolic function.

355

Factors determining the nature of progression of discrete fixed subaortic stenosis

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Introduction: In discrete fixed subaortic stenosis, surgery is indicated when the systolic gradient (Gmax) between the left ventricle (LV) and the aorta exceed 50 mmHg or in the presence of significant aortic regurgitation (AR).

The aim of this study is to determine the factors that influence the progression of the obstruction and the appearance of AR.

Methods: retrospective serial echocardiographic review of 19 patients, mean age 16 years (2 years-38 years), with fixed discrete subaortic stenosis that don't require surgery (initial Gmax at inclusion <50 mmHg and without any symptom). The mean follow up was 5.42 years. The progression of gradient is defined by the formula (Gmax at follow up – initial Gmax)

Results: the mean velocity of increasing of Gmax was 2 mmHg/year. This progression was correlated to the patient's age (cut off $=15$ years, $r=-0.5$; $p=0.02$), and the initial value of the Gmax (cut off $=40$ mmHg, $r=0.43$; $p=0.04$).

The appearance or the aggravation of AR was determined by: the initial grade of AR ($r=0.64$; $p=0.003$), initial Gmax ($r=0.65$; $p=0.002$), progression's velocity of G max ($r=0.47$; $p=0.04$), and distance between the membrane and the aortic cusps (cut off $=5$ mm, $r=0.49$; $p=0.03$). LV hypertrophy was influenced by the velocity of progression of obstruction (>2 mmHg/year)

Conclusion: the identification of factors determining the evolution of discrete subaortic stenosis (age <15 years, initial Gmax >40 mmHg, distance membrane-cusps >5 mm) allows an adequate screening of patients that will require early operation.

356

Ebstein's disease in adulthood

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Introduction: Ebstein's disease is a rare congenital malformation with many clinical manifestations depending on the age at diagnosis and the severity of the anatomical pattern. In this study we try to describe the clinical manifestations, the echocardiographic patterns and the outcome of this malformation in adulthood.

Methods and results: Retrospective study about 8 cases of Ebstein's anomaly in adult patients (5 women and 3 men); the mean age is 34 years old. The clinical manifestations were absent in 6% of cases but the majority of patients have exertional dyspnea or palpitations in 56% of cases. Arrhythmias and congestive heart failure were observed respectively in 6% and 13% of cases. Transthoracic echocardiography demonstrated the presence of Ebstein anomaly in all cases, showing apical displacement of the septal leaflet of the tricuspid valve. Five patients had the A grade of the Carpentier's classification and the others had the grade B. After an outcome of 11 years, one patient died by refractory heart failure.

Conclusion: Clinical manifestations and outcome of Ebstein's anomaly depend on the degree of tricuspid valve malformation and many cases are discovered in adults. The prognosis is worsening by arrhythmia and heart failure. Surgical intervention with tricuspid valve repair should be proposed for patients with severe heart failure and intractable arrhythmia. Conservative surgery and anti arrhythmic therapy had enhanced the prognosis of this congenital malformation.